On-line Supplement

eFigure 1. A summary of randomized controlled trials testing the effect of prostacyclin replacement therapy, phosphodiesterase type-V inhibitors, and endothelin receptor antagonists on mortality in PAH. Meta-analysis of published randomized controlled studies (identified by first author and year of publication) in pulmonary arterial hypertension (PAH) as of May 2010. The primary analysis has included 3780 patients of 23 trials. The figure shows the cumulative relative risk (RR) estimate of death in active treatment groups when compared with control groups stratified according to treatment class (inverse variance method). An overall reduction of mortality of 44% ($P = 0.016$) is shown. The subgroup analysis of the three classes of approved drugs (i.e., not including thromboxane receptor modulators) achieved a similar favorable reduction in mortality, although no statistical significance was achieved individually. Modified from Galiè N et al.[2]